



## Appendix A

### Claim Amendments

1. (Currently amended) A method for ~~preventing and/or~~ treating a respiratory disease in a patient comprising administering a therapeutically effective amount of a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~ to a patient in need thereof.
2. (Previously presented) The method as claimed in claim 1, wherein the respiratory disease is selected from the group consisting of COPD, bronchial asthma, pulmonary fibroses, emphysema, interstitial pulmonary disorders and pneumonias.
3. (Previously presented) The method as claimed in claim 1, wherein the respiratory disease is COPD.
4. (Currently amended) A method for ~~preventing and/or~~ treating muscular dysfunction in a COPD patient comprising administering a therapeutically effective amount of a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~ to a patient in need thereof.

5. (Currently amended) The method according to claim 1, wherein in addition to the compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~, a further compound selected from the group consisting of arginine, L-arginine hydrochloride (L-Arg HCl), L-arginine acetylaspariginat, L-arginine aspartate, L-arginine citrate, L-arginine glutamate, L-arginine oxoglurate, L-arginine tidiacicate and L-arginine timonacicate ~~or a derivative thereof~~ is used in simultaneous, separate or sequential combination with the compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~.
6. (Currently amended) A method for ~~preventing and/or~~ treating COPD in a patient comprising the step of administering a therapeutically effective amount of ~~BH4 or a derivative thereof~~ a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds to a patient in need thereof.

7. (Currently amended) The method according to claim 6, characterized in that in addition to a therapeutically effective amount of ~~BH4 or a derivative thereof~~ the compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds, a therapeutically effective amount of a further compound selected from the group consisting of arginine, L-arginine hydrochloride (L-Arg·HCl), L-arginine acetylaspariginate, L-arginine aspartate, L-arginine citrate, L-arginine glutamate, L-arginine oxoglurate, L-arginine tidiacicate and L-arginine timonacicate ~~or a derivative thereof~~ is administered in a simultaneous, separate or sequential combination with the compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~.
8. (Canceled)
9. (Canceled)
10. (Currently amended) A commercial product comprising: a customary secondary packaging, a primary packaging comprising a pharmaceutical preparation of a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of

these compounds ~~BH4 or a derivative thereof~~ and, optionally, a package insert, the pharmaceutical preparation being suitable for ~~prevention and/or~~ treatment of COPD in patients in need thereof.

11. (Currently amended) A method for ~~preventing and/or~~ treating a respiratory disease in a patient comprising administering a therapeutically effective amount of a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~ in combination with a therapeutically effective amount of a further compound selected from the group consisting of arginine, L-arginine hydrochloride (L-Arg·HCl), L-arginine acetylaspariginate, L-arginine aspartate, L-arginine citrate, L-arginine glutamate, L-arginine oxoglurate, L-arginine tidiacicate and L-arginine timonacicate ~~or a derivative thereof~~.
12. (Previously presented) The method as claimed in claim 11, wherein the respiratory disease is selected from the group consisting of COPD, bronchial asthma, pulmonary fibroses, emphysema, interstitial pulmonary disorders and pneumonias.
13. (Previously presented) The method as claimed in claim 11, wherein the respiratory disease is COPD.
14. (Currently amended) A method for ~~preventing and/or~~ treating muscular dysfunction in a COPD patient comprising administering a therapeutically effective amount of a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-

methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~, in combination with a therapeutically effective amount of a further compound selected from the group consisting of arginine, L-arginine hydrochloride (L-Arg·HCl), L-arginine acetylaspariginate, L-arginine aspartate, L-arginine citrate, L-arginine glutamate, L-arginine oxoglurate, L-arginine tidiacicate and L-arginine timonacicate ~~or a derivative thereof~~, to a patient in need thereof.

15. (Currently amended) A method for ~~preventing and/or~~ treating COPD in a patient comprising the step of administering a therapeutically effective amount of a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~ in combination with a therapeutically effective amount of a further compound selected from the group consisting of arginine, L-arginine hydrochloride (L-Arg·HCl), L-arginine acetylaspariginate, L-arginine aspartate, L-arginine citrate, L-arginine glutamate, L-arginine oxoglurate, L-arginine tidiacicate and L-arginine timonacicate ~~or a derivative thereof~~.
16. (Currently amended) A preparation comprising a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~ and a further compound selected from the group consisting of

arginine, L-arginine hydrochloride (L-Arg HCl), L-arginine acetylaspariginate, L-arginine aspartate, L-arginine citrate, L-arginine glutamate, L-arginine oxoglurate, L-arginine tidiacicate and L-arginine timonacicate ~~or a derivative thereof~~ as a combined preparation for simultaneous, separate or sequential administration.

17. (Currently amended) A pharmaceutical composition comprising a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~ and a further compound selected from the group consisting of arginine, L-arginine hydrochloride (L-Arg HCl), L-arginine acetylaspariginate, L-arginine aspartate, L-arginine citrate, L-arginine glutamate, L-arginine oxoglurate, L-arginine tidiacicate and L-arginine timonacicate ~~or a derivative thereof~~.
18. (Previously presented) The pharmaceutical composition as claimed in claim 17 further comprising a pharmaceutically acceptable carrier.
19. – 22. (Canceled)
23. (Currently amended) A trade package comprising as pharmaceutical agent a compound selected from the group consisting of (6R)-L-erythro-5,6,7,8-tetrahydrobiopterin (BH4), (6R,S)-5,6,7,8-tetrahydrobiopterin, 1',2'-diacetyl-5,6,7,8-tetrahydrobiopterin, Sepiapterin, 6-methyl-5,6,7,8-tetrahydrobiopterin, 6-hydroxymethyl-5,6,7,8-tetrahydrobiopterin, 6-phenyl-5,6,7,8-tetrahydrobiopterin, and the pharmaceutically acceptable salts of these compounds ~~BH4 or a derivative thereof~~ and/or a compound selected from the group consisting of arginine, L-arginine hydrochloride (L-Arg HCl), L-

arginine acetylaspariginate, L-arginine aspartate, L-arginine citrate, L-arginine glutamate, L-arginine oxoglurate, L-arginine tidiacicate and L-arginine timonacicate ~~or a derivative thereof~~ together with instructions for use of the pharmaceutical agents in combination for simultaneous, separate or sequential administration for the ~~prevention and/or~~ treatment of respiratory diseases.